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CRAO And CRVO: Separate Paths To A Familiar Impasse

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Abstract

We aim to discuss the proceedings of a rare case of Combined Central Retinal Artery Occlusion (CRAO) and Central Retinal Vein Occlusion (CRVO) with Cerebral Venous Sinus Thrombosis (CVST). A 28-year-old male diagnosed with thrombosis of transverse and sigmoid sinus was referred to us with sudden bilateral diminution of vision for five days. Detailed Ophthalmic examination revealed a vision of perception of light (PL) bilaterally which decreased to a complete loss of vision in both eyes within one week. Fundus examination bilaterally revealed multiple intraretinal hemorrhages with occluded vessel in background of a pale retina. Diagnosis of CRAO with CRVO was made. Patient was put on antiepileptic and anticoagulant along with steroids to which he responded systemically. We present a rare case of a combined diagnosis of CRVO and CRAO with cerebral venous sinus thrombosis

Key Message: Combined presentation of CRAO and CRVO in a male with cerebral venous thrombosis is a rare presentation which points to a deeper systemic disease along with poor visual prognosis

Keywords: Combined Central Retinal Artery and Vein Occlusion, Systemic venous thrombosis, hypercoagulable disorders

Introduction

A review of the literature on the rare cases of combined Central Retinal Artery Occlusion (CRAO) and Central Retinal Vein Occlusion (CRVO) have revealed that common etiological risk factors for both the discrete pathologies lead to their co-existence. [1 -8] There are different types of combination where the patient can either have a CRVO with CRAO, CRVO with Branch Retinal Arterial Occlusion (BRAO) and CRVO with cilioretinal artery occlusion (CLRAO). [1] Vasculitis and thromboembolic disorders are the main differential diagnosis for a combined CRAO and CRVO pathology, especially in the younger age group. [2] Cilioretinal artery block is not caused by embolic event but rather by hemodynamic block. [1] The diagnosis of a combined retinal artery and venous occlusion is based on the presence of sudden loss of vision with retinal oedema along with haemorrhages, with or without a cherry red, venular tortuosity and engorgement, delayed arterial filing and prolonged arteriovenous transit time in fluorescein angiography. [1 - 7] The incidence of CVST in young adults is three cases per million per year with predilection to female gender making our case a rare phenomenon. Common risk factor for retinal vascular occlusion and Cerebral Venous Sinus Thrombosis (CVST) in our case seemed to be a hypercoagulable disorder of unknown origin.

Case Report:

A 28-year-old male was referred to us with sudden, painless and progressive diminution of vision for five

days in both eyes. There was no history of any similar episodes in the past. Patient did not have any underlying systemic illness at the time of presentation. MRI done outside suggested a thrombosis in transverse and sigmoid sinus and the patient was admitted under the neurosurgery department of the hospital.

On examination patient had only perception of light in both eyes which was accurate in all four quadrants. Bilateral eye pupil was four mm dilated and sluggishly reacting to light. Rest of the anterior segment findings were within normal limits. Extraocular movements were free and full in all gazes and direction and IOP was within normal limits. Fundus evaluation of both eyes revealed extensive multiple intraretinal superficial and deep haemorrhages with attenuated white vessels noted in the posterior pole and peripapapillary area in all four quadrants extending to the periphery along with tortuous and engorged veins suggesting a CRVO. These findings were present in the background of a pale retina in macular area and the presence of attenuated arteries which suggested a diagnosis of CRAO. (Figure 1 and 2). There was no disc oedema in both the eyes. The patient was in altered sensorium and therefore fundus fluorescein angiography or other ancillary examination like OCT could not be done to confirm the diagnosis. Over the course of seven days, the patient had complete loss of vision in both eyes and also developed vitritis in Right Eye.

Haematological investigations were done to reach a systemic diagnosis. A complete blood count along with peripheral smear showed microcytic, hypochromic anaemia (Hb = 8.20 gm %). PT INR values were normal and ESR was raised (18 mm/hr). 2D echo, renal and liver function tests were within normal limits. CT orbit scan was normal. Patient was treated with Intravenous Methylprednisolone one mg/kg body weight for five days followed by oral tapering dose along with IV Levipil (antiepileptics) and Injection Clexane (anticoagulants) for 10 days followed by oral long term warfarin. Patient was also referred to a Neurophysician who advised detailed blood tests to rule out inherited hypercoagulable disorder but any further investigations for a definitive diagnosis was not consented by the patient and his relatives. Patient was discharged after significant systemic improvement though vision did not improve.

Discussion:

Pathogenesis of combined presentation of CRAO and CRVO is till date unclear. [9] CRVO has been postulated to be the initial event where a sudden rise in the venous pressure which when exceeds the systolic BP causes compromise in the arterial circulation due to back pressure. [7] CLRAO is usually present secondary to CRVO due to the hemodynamic block created by the distended and engorged CRV which probably was the reason for complete loss of vision by the end of 7 days after the patient presented to us. [1]

CVST is a very rare form of venous thromboembolism which is found predominantly in younger population. 90 % of the time it presents at multiple location like sigmoid and transverse sinus like in our case. [8] Females are more prone with documented F: M ratio of 3: 1. CVST is multifactorial and is caused by either inherited or acquired risk factors. In 20 – 35 % of the cases the cause cannot be identified. Hereditary risk factors involve homocysteinemia, factor V Leiden homozygous mutation, protein C and S and anti-thrombin III deficiency, and positive anticardiolipin or antiphospholipid antibodies. On the other hand, acquired risk factors include brain tumours, head trauma, central nervous system infections, intracranial hypotension, extracerebral neoplasia, dural fistulas, haematological conditions, nephrotic syndrome, neurological surgery, lumbar puncture. [8]

The common pathogenesis for retinal vascular occlusion and CVST is implied by the classical Virchow's triad of thrombogenesis. The three known components being hypercoagulability, blood stasis and vessel wall damage. [1, 8] Hypercoagulable disorders have been known to cause combined CRVO and CRAO presentation by many others [2, 4, 5] and also justifies the cerebral venous thrombosis. Table 1 compares the baseline characteristics of our case report with previous studies.

The visual prognosis is very poor in cases of combined arterial and venous occlusions where the visual acuity is normally in hand movement range. [9] According to a study done by Schmidt D only five out of the 10 patients (50 %) with combined CRAO and CRVO improved in terms of visual acuity on treatment. This showed poor visual prognosis when compared to the cases of only CLRAO with CRVO

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where Eight of those nine eyes documented a final visual acuity of 20/30 or better. Coppeto & Lessell have documented total bilateral blindness where arterial and venous obstruction coexisted. [1]

What makes our case unique is that the combined retinal vascular occlusion along with CVST, both being rare entities is present in a gender which has an uncommon predilection for the same. One case of septic cavernous sinus thrombosis has been documented along with combined retinal vascular occlusion [3] but none in the background of a hypercoagulable disorder.

Conclusion:

We present a previously undocumented case where a retinal vascular occlusion is present along with cerebral venous sinus thrombosis in background of undiagnosed hypercoagulable disorder. A combined diagnosis of central retinal arterial and venous obstruction comes with a very poor visual prognosis regardless of the primary cause and the treatment given to the patient. In such cases, the importance of life overpowers the visual outcome and the same needs to be counselled to the patient and his/her well-wishers.

Declaration Of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms for usage of clinical information and images to be reported in the journal. The patient understand that their personal information will not be revealed in the article but cannot be guaranteed.

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TABLE 1: Comparison of baseline characteristics of our case with previous studies

STUDY	SAMPLE SIZE	F/M RATIO	MEAN AGE (Years)	DIAGNOSIS	ETIOLOGY

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Schmidt et al [1]	14	5:9	57.6	10 – CRVO + CRAO 3 – CRVO + BRAO 1 – CRAO + CRVO	8 – Arterial HTN 3 – hypercholestremia 6 – Coagulation Anomaly
Lemos JA [2]	1 (Female)	-	49	CRAO + CRVO	Factor V laiden mutation
Richards RD [3]	6	1:1	25	CRAO+CRVO	Behcet's Disease
			48	CRAO+CRVO	Vasculitis
			36	CRAO+CRVO	Septic cavernous sinus thrombosis
			52	CRAO+CRVO	Subacute bacterial endocarditis
			51	CRAO+CRVO	Lymphocytic lymphoma
			38	CRAO+CRVO	Acute lymphocytic leukemia
Chakravarthi MD [4]	1(male)	-	42	CRAO+CRVO	Protein C deficiency
Desai S [5]	1 (male)	-	33	CRAO+CRVO	Type 2 protein C deficiency with factor V leiden mutation
Parchand SM [(6)]	1 (female)	-	16	CRAO +CRVO	SLE
Current case	1(male)	-	28	CRAO+CRVO+CVTS	Unknown

FIGURE 1 - RE showing multiple retinal haemorrhages with thin attenuated arteries with retinal whitening in macular area

FIGURE 2 - LE showing multiple retinal haemorrhages with vascular beading noted with retinal whitening in macular area

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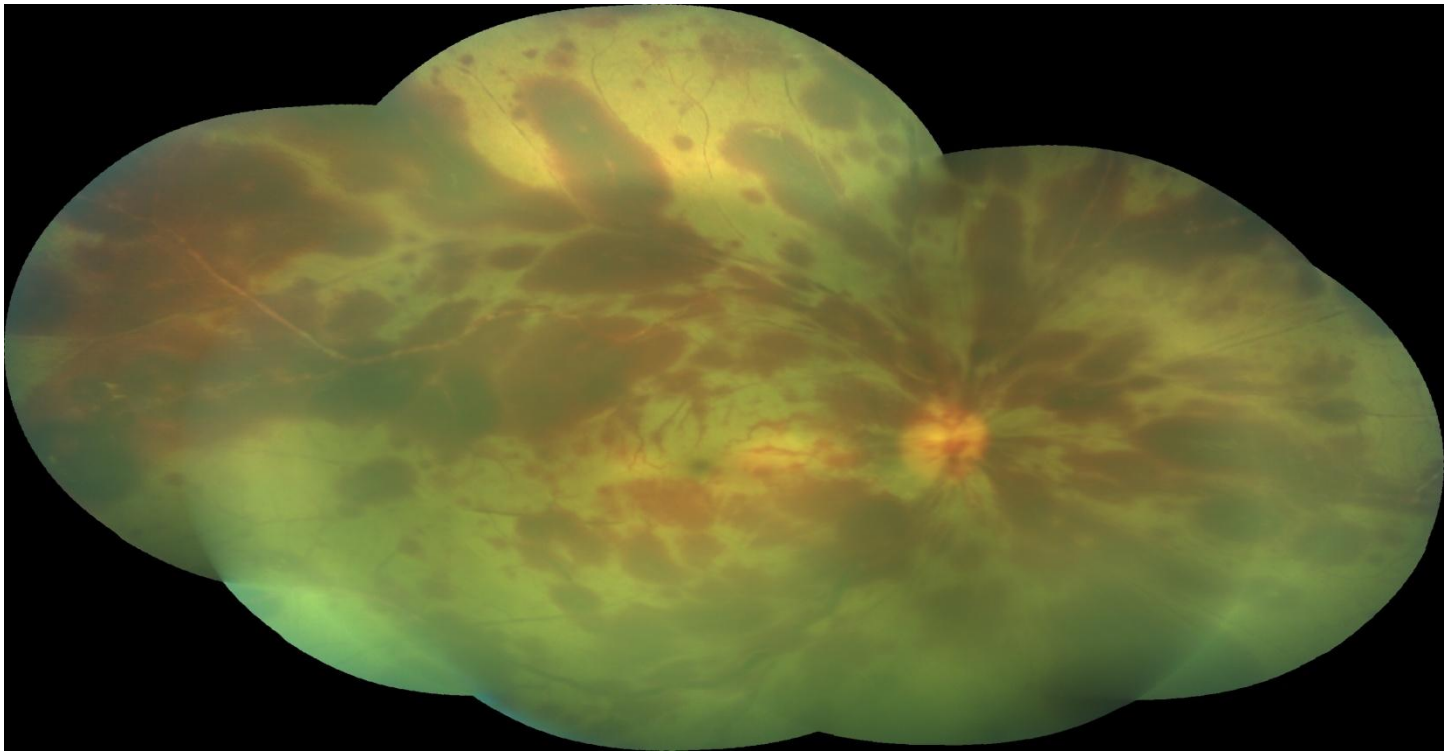

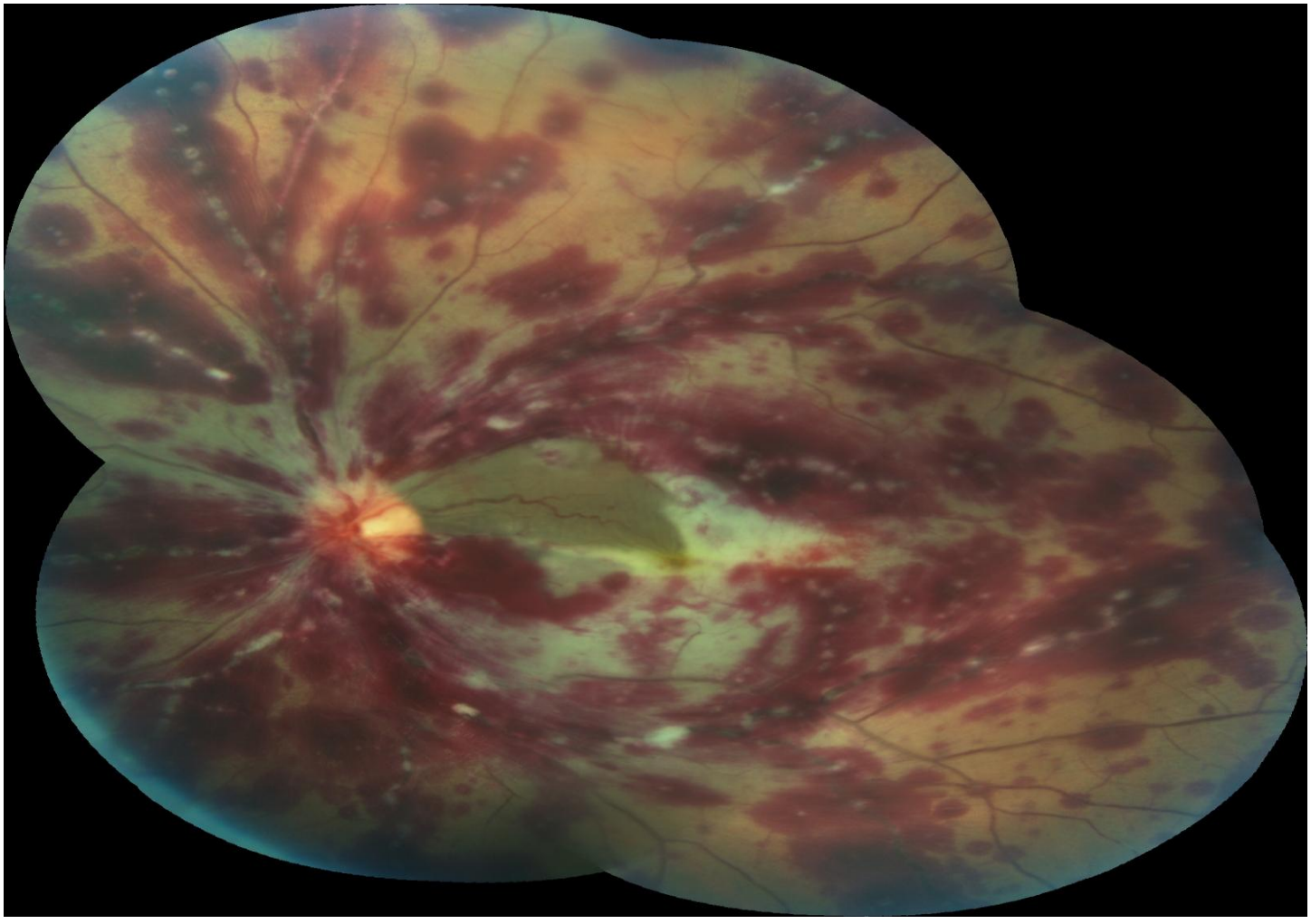

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